

Twenty-Three-Year Follow-up of Separated Ischiopagus Tetrapus Conjoined Twins

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This paper presents a 23-year follow-up of the separation of ischiopagus tetrapus conjoined twins reported in *Annals of Surgery* in December 1966. One twin died of septicemia at age 2 years after bilateral pelvic osteotomies for the treatment of her marked pelvic diastasis. The surviving twin has done reasonably well, and her most significant problem is related to her musculoskeletal system. She has an increasing T7-10, L-1 apex right congenital scoliosis with wedged vertebra at T-10, as well as marked pubic diastasis and bilateral subluxation of her hips. This has resulted in a somewhat aberrant physical appearance and a "waddling" gait. Her colostomy functions well and she has normal renal and bladder function. This patient's history illustrates that many problems remain after successful separation of conjoined twins. However these problems are manageable and do not preclude the possibility that such a patient may be a productive member of society.

THERE ARE MORE THAN 600 publications in the literature concerning conjoined twins and they treat many topics, including prenatal diagnosis, obstetrical management, labor and delivery, neonatal care, preoperative evaluation, anesthetic management, and various aspects of surgical separation.¹⁻¹⁵ However there have been few reports on the long-term follow-up of conjoined twins after surgical separation. This paper reports on the follow-up of ischiopagus tetrapus twins separated at our institution and reported in *Annals of Surgery* in 1966, with a review of the literature on the separation of all ischiopagus conjoined twins.

The incidence of conjoined twins is approximately 1 in 25,000 to 1 in 80,000 births, with most authors quoting an average incidence of 1 in 50,000 births.^{6,14,16} However

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EVALUATION OF ISCHIOFAGUS TETRAPUS



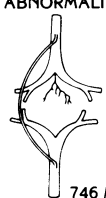
SYSTEM	STUDY	FINDINGS
• UROGENITAL TRACT INNervation	• I.V.P. RENAL SCAN • CYSTOMETROGRAMS BEFORE & AFTER SPINAL BLOCK • VAGINOGRAM	
• ALIMENTARY TRACT	• UPPER GI SERIES • SMALL BOWEL FOLLOW THROUGH • BARIUM ENEMA	
• CARDIORESPIRATORY	• CHEST FILMS • EKG	NO ABNORMALITIES
• VASCULAR SYSTEM	• RETROGRADE BRACHIAL AORTOGRAMS	
• BLOOD VOLUME & SHUNT	• RADIOISOTOPE STUDY WITH LABELLED ALBUMIN	746 ML. 19 ML./MIN.
• ENDOCRINE RELATIONSHIPS DEPENDENCY	• ¹³¹ I UPTAKE & SCAN	• BOTH TWINS HAD FUNCTIONING THYROID
• SKELETAL SYSTEM	• ROENTGENOGRAMS	• SCOLIOSIS & SPINA BIFIDA

FIG. 1. Preoperative evaluation of potentially joined or dependent organ systems.

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TABLE 1. *Surgical Separation of Conjoined Twins*

Year	Author and Reference	Sex	Type	Age at Separation	Shared Organs	Indication of Separation	Results (Follow-up)	
							Twin 1	Twin 2
1955	Spencer ^{18,22}	F	Isch Tet	18 hrs.	Liver bridge, terminal ileum, colon, anus, pelvic organs	Emergent; demise of twin one	D (sacrificed)	S (age 10 mos.)
1966	Eades ¹⁹	F	Isch Tet	7 mos.	Colon, anus, pelvic organs	Elective	S (age 23 yrs.)	S (died at 2 years)
1967	Akiyama ²⁴	F	Isch NS	3 mos.	NS	NS	S	D
1968	Mestle ^{21,23}	F	Isch Tri	16.5 mos.	Colon, anus	Elective	S (age 20 yrs.)	S (age 20 yrs.)
1969	Poradowsky ²⁶	F	Isch Tet	NS	Pelvic organs, cloaca	Emergent: intestinal obstruction	D	D
1969	Borden ²⁷	F	Isch Tet	NS	Ileum, colon, liver bridge	Elective	S	S
1969	Koop ²⁸	NS	Isch Tet	NS	NS	NS	S	S
1971	Rossi ²⁹	F	Isch Tet	NS	Terminal ileum, colon	Elective	S (age 2 yrs.)	S (age 2 yrs.)
1971	Borde ³⁰	F	Isch Tri	3 yrs.	Colon, rectum, liver bridge, bladders	Elective	D (10 hrs. postop.)	D (36 hrs. postop.)
1971	Mayell ³¹	NS	Isch Tri	NS	NS	NS	D	S
1971	Bankole ^{32,33}	F	Isch Tet	60 hrs.	Ileum, colon, bladder, cloaca	Emergent: intestinal and urethral obstruction	D (30 hrs. postop.; congestive heart failure)	D (congestive heart failure)
1971	Furman ³⁴	F	Isch Tet	4 days	Liver bridge, colon, anus, pelvic organs	Emergent: intestinal obstruction	S	S
1974	Cywes ³⁵	F	Isch Tet	7 days	Bladder, cloaca	Emergent: intestinal obstruction	D (6 days postop. sepsis)	D (sacrificed anencephaly)
1974	Koop ²⁸	NS	Isch Tet	NS	NS	NS	S	S
1976	Akiyama ²⁴	F	Isch Tet	3 mos.	Terminal ileum, colon, rectum, bladder, cloaca	Elective	S	S
1978	Rosenburg ^{20,21}	F	Isch Tet	13 mos.	Colon, anus, pelvic organs	Elective	S (age 13 yrs.)	D (1 yr. postop. aspiration)
1979	Hung ³⁶	M	Isch Tri	2.75 yrs.	Liver bridge, terminal ileum, colon, penis	Elective	S (age 9 yrs.)	S (age 9 yrs.)
1979	Chao ¹	M	Isch Tet	2.75 yrs.	Liver bridge, colon, anus, penis	Elective	S	S
1981	Rector ³⁷	F	Isch Br	11 mos.	Colon, rectum, bladder, liver bridge	Elective	S (age 8 yrs.)	S (age 8 yrs.)

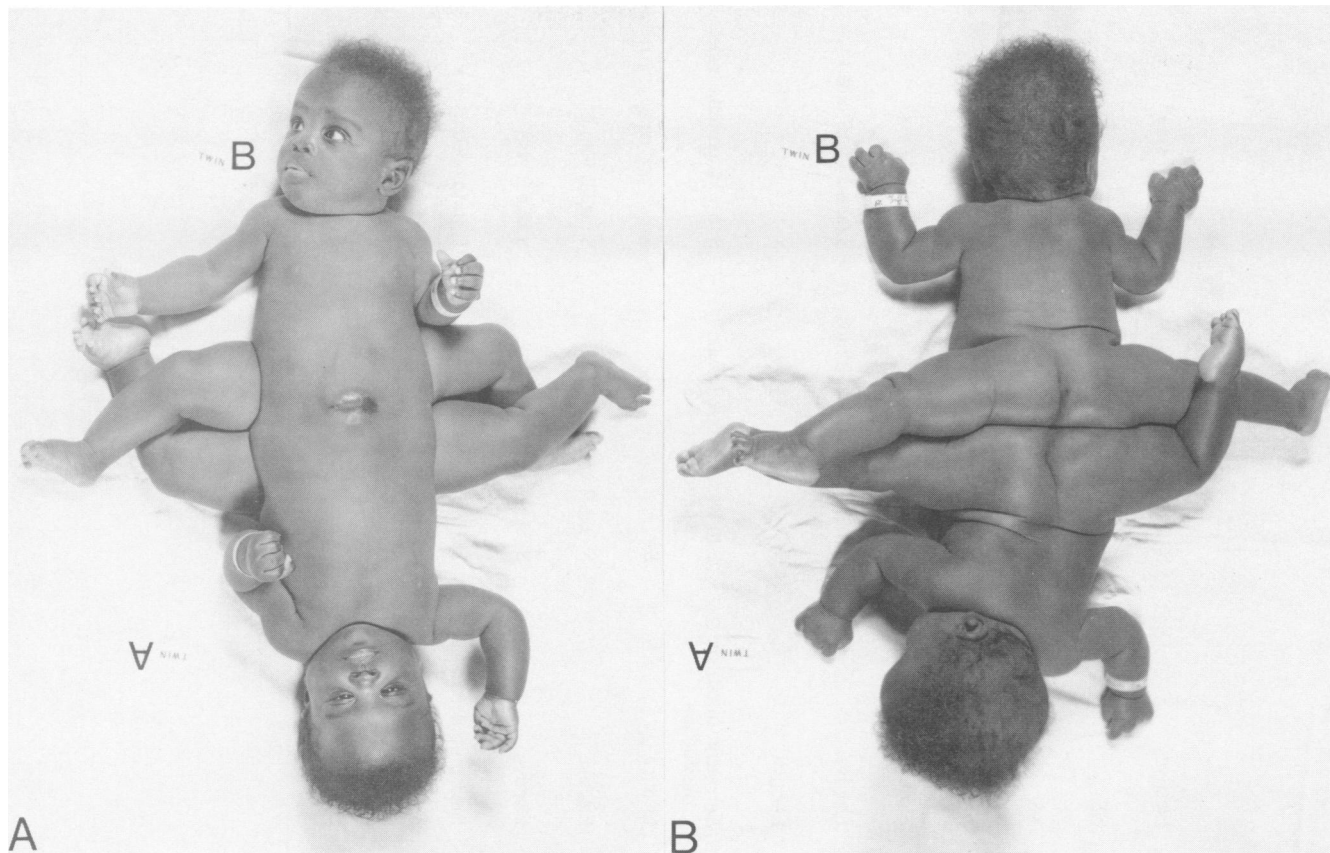
1981	Somasundaram ³⁸	F	Isch Tet	7 mos.	Colon	Elective	S (age 4.5 yrs.)	S (age 4.5 yrs.)
1981	Vottler ⁸	M	Isch Tri	NS	NS	NS	S	S
1981	Vottler ⁸	F	Isch Tri	NS	NS	NS	S	D
1981	Vottler ⁸	F	Isch Tri	NS	NS	NS	S	S
1982	Somasundaram ³⁸	F	Isch Tet	4 mos.	Colon, urethra	Elective	S (age 3.25 yrs.)	S (age 3.25 yrs.)
1982	Grantzow ³⁹	M	Isch Tri	2.5 yrs.	Terminal ileum, colon, pelvic organs, liver	Elective	S (died at age 3 yrs. 8 mos.; sepsis)	S (age 3 yrs. 8 mos.)
1983	Grantzow ⁴⁰	M	Isch Tet	10 mos.	Terminal ileum, colon, pelvic organs	Elective	S	S
1984	Harrison ⁹	M	Isch Tet	4 days	Colon, Penis, bladder	Emergent; intestinal obstruction	S (6 wks. postop.; female assignment)	S (6 wks. postop.; male)
1984	Savickis ⁴¹	M	Isch Tri	2.5 yrs.	Liver bridge, colon, bladder, penis	Elective	S	S
1984	James ²	M	Isch Tri	29 mos.	Colon, liver, penis, prostate	Elective	S (age 2 yrs.)	S (age 2 yrs.)
1984	Ross ³ , O'Neill ⁴²	F	Isch Tri	4 mos.	Liver bridge, colon, terminal ileum	Elective	S (age 2 yrs.)	S (age 2 yrs.)
1984	Olcay ¹⁶	M	Isch Tet	? days	Pelvic organs	Emergent: demise of twin one	D (14 hrs. postop.)	D (intraoperatively)
1985	Zuker ⁴³	M	Isch Tet	9 mos.	Colon, penis	Elective	S (3 mos. postop.)	S (3 mos. postop.)
1986	Chatterjee ⁴⁴	F	Isch Tet	1 yr.	Colon, anus, pelvic organs	Elective	S (6 mos. postop.)	S (6 mos. postop.)

Isch, ischiopagus; Tet, tetrapus; Tri, tripus; Bi, bipus; F, female; M, male; NS, not stated;

S, survived; D, died.

Pelvic organs are as described in the text.

Listed year is year of separation or year of publication.



FIGS. 2A and B. (A) Front and (B) posterior view of ischiopagus tetrapus twins at 7 months of age.

60% are stillborn, yielding an incidence of conjoined twins alive at birth of 1 in 200,000. In Africa the incidence has been quoted as high as 1 in 14,000 deliveries, thus suggesting an increased frequency in blacks.

Conjoined twins are classified by their most prominent site of connection with the Greek root *pagus* (that which is fixed). Such sites may include the thorax (thoracopagus), abdomen (omphalopagus), pelvis (pyopagus), or skull (craniopagus). Thoracopagus is the most common classification, accounting for 40% of reported cases; omphalopagus number 33%; pyopagus, 19%; ischiopagus, 6%; and craniopagus, 2%.^{8,17} Ischiopagus twins are joined at the pelvis and they usually have three (tripus) or four limbs (tetrapus). The intestinal tracts usually join at the terminal ileum, which empties into a single colon. Four kidneys and two bladders are usually present. One ureter from each twin frequently crosses the midline and empties into the bladder of the other. Male twins may have fusion of the genitourinary tract distal to the bladder but they usually have four gonads. Female twins usually have four ovaries, two uteri, and two sets of external genitalia on either side of a single anus. Vaginal anomalies and rectovaginal communication are not uncommon.

To date there have been 33 attempts at surgical separation of ischiopagus conjoined twins (Table 1). The first separation of ischiopagus twins with survival of one twin was performed by Spencer¹⁸ in 1955. In 1966 Eades and Thomas¹⁹ reported the first separation of ischiopagus twins with survival of both twins.

We report a 23-year follow-up of the surviving twin.

Case Report

The twins were born April 6, 1965 and transferred to the North Carolina Memorial Hospital. Studies revealed the following (Fig. 1): (A) A widely open, semicircular pelvis with fusion of each pubic symphysis with the corresponding symphysis of the opposing twin. (B) Thoracolumbar scoliosis and spina bifida involving the lower lumbar vertebrae in both twins. (C) A common distal ileum, cecum, and colon terminating in a rectal fistula. (D) Two laterally placed bladders receiving ureters from both twins with bilateral innervation of both bladders. (E) Bilateral arterial supply to the pelvic organs and a direct continuation of the superior mesenteric artery of infant A to infant B. Bilateral blood supply to the common colon from the inferior mesenteric artery of each twin. (F) Absence of kidney and ureter on the left side of infant A.

The twins were separated at age 7 months on November 8, 1965 (Fig. 2). The union between the bony pelvis of the infants was separated. Each infant was provided with one kidney, ureter, and bladder, essentially complete internal genitalia, a portion of colon, and a small intestine of

normal length. Both twins were given a permanent colostomy in the right midabdomen. At the time of publication (4 months after operation), the twins were doing well and undergoing progressive splinting to restore a more normal position at the hips and permit future ambulation because both twins had a marked pubic diastasis and bilateral coxa vara.

Twin A's postoperative course was complicated by the development of phlegmasia cerulea dolens secondary to thrombophlebitis caused by a venous catheter high in her right axillary vein. She subsequently developed ischemic necrosis of skin on her distal right forearm requiring multiple debridements and skin grafts. This progressed to chronic osteomyelitis of her right distal ulna and radius. At approximately 3 years of age and two days after undergoing bilateral pelvic osteotomies for treatment of marked pubic diastasis, she died of cardiac arrest. Autopsy revealed streptococcal septicemia, principally in her lungs and brain.

Twin B's most significant problem is related to her musculoskeletal system; she has an increasing T7-10, L-1 apex right congenital scoliosis with wedged vertebrae at T-10, as well as marked pubic diastasis and bilateral subluxation of her hips. Recommendation for surgical amelioration of these problems was not accepted by the patients family. At age 4 years her scoliosis measured 75 degrees; at age 6 years it measured 108 degrees; and at age 20 years it was 107 degrees (Fig. 3). She continues to have a widely separated pubic symphysis (Fig. 4). She also has a marked kyphosis of 83 degrees.

Menarche occurred at approximately age 12 to 13. In 1981 at age 16, she underwent a right salpingo-oophorectomy for removal of a symptomatic (painful) right ovarian cyst measuring 8 cm in diameter. Pathologic findings were those of endometriosis with an endometrial cyst. She has severe lower abdominal pain in association with menses such that she must remain in bed for two to four days. This started in 1985 and is still a recurring problem. The overall findings are consistent with endometriosis and she has been treated with oral contraceptive therapy.

She has always been a bright student and maintained a B average in the 8th grade; however she dropped out of school in the 11th grade. She indicates that this was because of a fall that resulted in discomfort in her left hip such that she could not walk more than one block without stopping.

Her mother died in 1984 of pneumonia and because she is the oldest in the household she assumed the responsibility of the household for herself and her two brothers, aged 16 and 10 years. Her grandmother and aunt live nearby.

Twin B is now 23 years old and was last seen in 1985 with full body photos (Fig. 5).

Discussion

Of 33 attempts at surgical separation of ischiopagus conjoined twins, 11 were ischiopagus tripus and 20 were ischiopagus tetrapus (Table 1). One case was not declared as tripus or tetrapus and another was ischiopagus bipus (sharing only two legs). Sixty-seven per cent of these twins were female (tetrapus, 70%; tripus, 45%). Shared organs included a common terminal ileum in 27.3% (9 cases) and a colon in 69.7% (23 cases). The perioperative mortality rate for ischiopagus conjoined twins was 18.8%, with 52 of 66 twins living (excluding two twins who had to be sacrificed to save their conjoined siblings). There was no difference in perioperative mortality rate between ischiopagus tetrapus and tripus twins. Before 1975 the perioperative mortality rate was 38.46% (16 of 26 twins living) and from 1975 through 1987 it was reduced to 10% (36

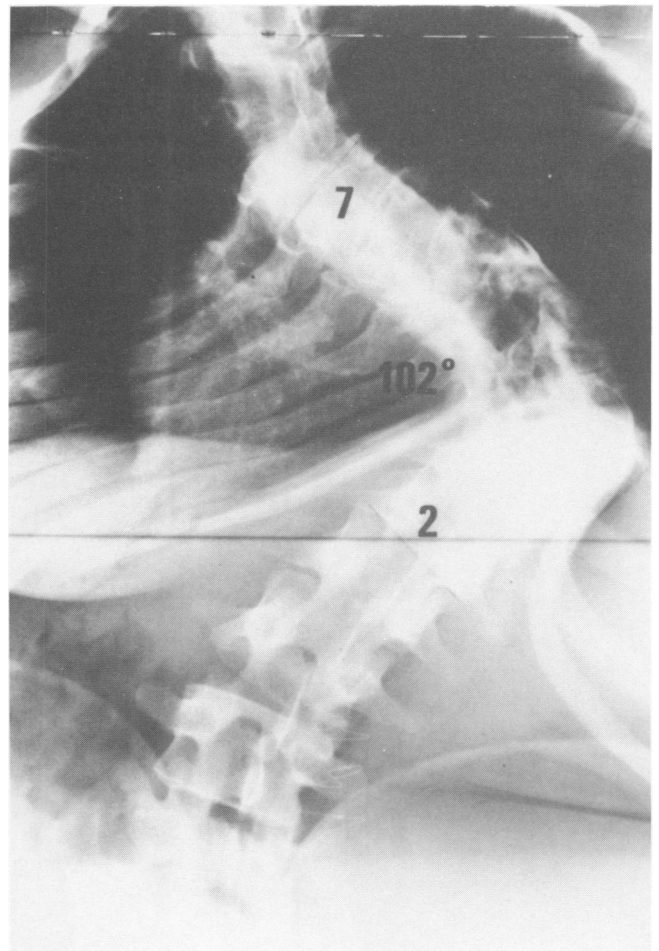


FIG. 3. Radiograph demonstrating scoliosis of 107 degrees at age 21 years.

of 40 twins living). Analysis of the effect of emergent operation on outcome revealed an overall perioperative mortality rate of 18.8% compared to 64.3% with emergent operation and 5.26% with elective operation.

Reports of the follow-up of surgically separated twins are few. Rosenberg²⁰ reported the successful separation of ischiopagus tetrapus twins in 1978 with the survival of both twins. One twin died 1 year after operation from aspiration of a bean, the other has been partially deaf since birth, has had problems with urinary tract infections and hematuria, has a somewhat disturbed gait as a result of pubic diastasis, but otherwise is a healthy, sixteen year old.²¹

Spencer¹⁸ reported the successful separation of ischiopagus tetrapus twins in 1955. One twin was deformed and moribund. The other twin was lost to follow-up at 10 years of age but at that time was doing quite well except for her short stature and abnormal gait due to the absence of symphysis pubis.²² Mestel²³ reports ischiopagus tripus

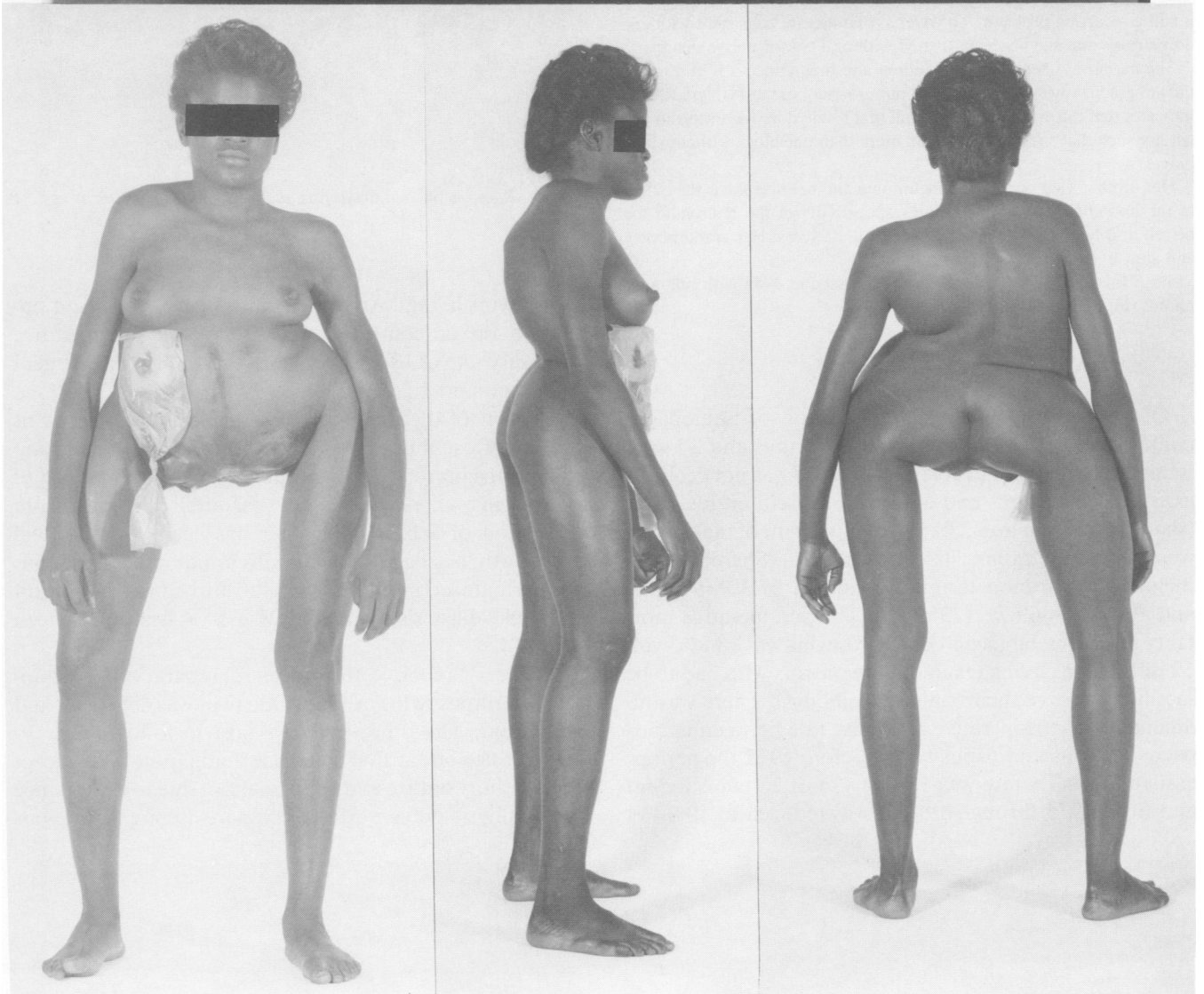
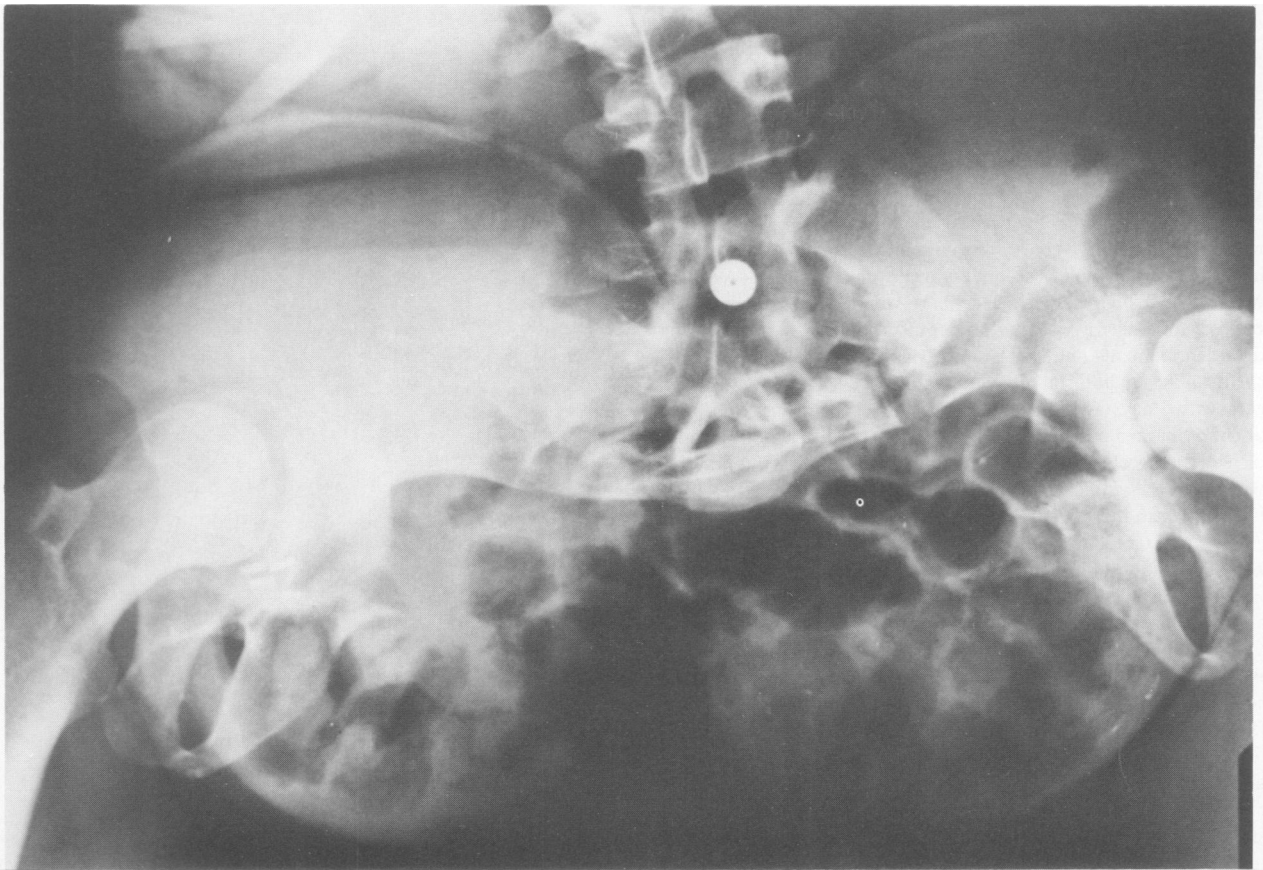


FIG. 4. Radiograph demonstrating widely separated pubic symphysis at age 20 years.

FIGS. 5A–C. (A) Front, (B) lateral, and (C) posterior appearance at age 20 years.

twins who are functioning with a prosthesis and doing quite well 20 years after surgical separation.

Successfully separated ischiopagus twins require long-term rehabilitation and follow-up, particularly with regards to orthopaedic, urologic, and gynecologic problems. However, despite all these problems, separation can enable these patients to become happy, intelligent, and potentially productive members of society.

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